EDITORIAL

Sudden cardiac death in young athletes

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Most cases of sudden cardiac death in young athletes (<35 years) are caused by inherited cardiomyopathies, notably hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy

Regular exercise reduces the risk of atherosclerosis and subsequent sudden cardiac death (SCD).[1] In this regard, athletes are perceived as the epitome of health, owing to their unique lifestyle and extraordinary physical achievements. However a small, but notable proportion of athletes die suddenly.[2] Such tragedies are highly publicised, particularly when high-profile athletes are involved. In the past year, the UK has witnessed seven highly publicised deaths during sport between the Great North run, London Marathon and London Triathlon, which have caused major concern among sporting bodies, doctors and the lay public. Most of the deaths in athletes are due to disorders of the cardiovascular system. Coronary artery disease is the most common cause of SCD in athletes aged >35 years. However, in younger athletes most SCDs are attributed to inherited or congenital disorders of the heart that predispose to malignant ventricular arrhythmias.[2 3] We focus on young (age <35 years) athletes.

MAGNITUDE OF THE PROBLEM

The precise incidence of SCD in athletes is unknown, but estimates from the USA suggest a figure between 1 in 200 000 and 1 in 300 000.[4] This figure is likely to be an underestimate for three important reasons. Firstly, there is no systemic national registry for sudden death in sports, and data are derived from high-profile events or deaths of prolific athletes. Secondly, an expert cardiac pathologist is rarely responsible for conducting postmortem examinations on athletes, and relatively rare conditions such as arrhythmogenic right ventricular cardiomyopathy (ARVC) or atypical forms of the more common conditions such as hypertrophic cardiomyopathy (HCM) may not be identified. Finally, deaths from ion channelopathies or accessory electrical pathways are not identified during a postmortem examination.[5]

Most data regarding the demographics of athletes dying suddenly are derived from a large American series of 158 athletes.[6] The study showed that most deaths occur in adolescent males (mean age 17.1 years), with a higher preponderance in black athletes. Most deaths occurred during or just after physical exertion, suggesting that adrenergic surges promote fatal cardiac arrhythmias in patients harbouring serious cardiovascular disorders.

CARDIOVASCULAR CAUSES OF SUDDEN DEATHS IN ATHLETES

Most SCDs in athletes are due to inherited cardiomyopathies, notably HCM and ARVC.[2 3] In the American series, HCM is the most common culprit accounting for up to one third of all deaths; however, data from the Veneto region of Italy suggest that ARVC is the most common cause.[7] The aetiological difference may partly reflect the fact that the annual screening of all athletes in Italy selects out individuals with HCM. Coronary artery anomalies are relatively common and usually benign. The most serious and potentially fatal form comprises the left main coronary artery arising from the right sinus of Valsalva, resulting in acute leftward passage of the coronary artery along the aortic wall, causing the entrance to be slit-like.[8]

Although most individuals with cardiomyopathy are unable to exercise intensively, a small proportion are capable of participating in competitive sport at the national level. This is exemplified by the cases such as those of Mark Vivian Foe, the Cameroon football star who played for several seasons in the British premier league and died from HCM during an international game in 2003, as well as the death of a club runner in the London Marathon in 2005 who had run 19 marathons previously in < 3.5 h.

SCD in athletes may also occur from acquired causes, such as myocarditis, commotio cordis, drug misuse or trauma (box 1).

Approximately 2% of athletes who die suddenly do not have an identifiable cause at postmortem examination.[5 6] Experience from studies in the first-degree relatives of patients who are victims of sudden adult death syndrome indicates that a proportion of these deaths are attributable to ion channelopathies (congenital long QT syndrome (LQTS) and Brugada syndrome). Fatal tachyarrhythmias due to undiagnosed accessory pathways comprise other potential causes (box 2).

CARDIAC EVALUATION OF ATHLETES

The investigation of an athlete in the UK is most commonly part of a screening programme established by certain sporting bodies such as the British Lawn Tennis association, and premier rugby and football leagues. Rarely, cardiac evaluations may be triggered by cardiovascular symptoms reported by an athlete or because of the presence of

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Abbreviations: ARVC, arrhythmogenic right ventricular cardiomyopathy; HCM, hypertrophic cardiomyopathy; LQTS, long QT syndrome; LVH, left ventricular hypertrophy; SCD, sudden cardiac death

Box 1 Causes of sudden cardiac death in young athletes

Congenital/inherited causes

- Disease of the myocardium

  Hypertrophic cardiomyopathy
  Arrythmogenic ventricular cardiomyopathy
  Dilated cardiomyopathy

- Coronary artery disease/anomalies

  Anomalous insertion of coronary arteries
  Premature atheromatous coronary artery disease

- Cardiac conduction tissue abnormalities

  Wolff–Parkinson–White syndrome
  Right ventricular outflow tachycardia
  Catecholaminergic polymorphic tachycardia

- Valvular heart disease and disorders of the aorta

  Mitral valve prolapse
  Congenital aortic stenosis
  Marfan syndrome

- Ion channelopathies

  Congenital long QT syndrome
  Brugada syndrome

Acquired heart disease

- Infections (myocarditis)
- Drugs (cocaine, amphetamine)
- Infiltrative disorders (sarcoid, amyloid)
- Electrolyte disturbances (hypokalaemia or hyperkalaemia)
- Hypothermia
- Hyperthermia
- Trauma (commotio cordis)

a family history of premature cardiac disease and/or SCD in first-degree relatives. Ideally, athletes should be investigated in centres with expertise in the diagnosis and management of inherited cardiac disorders, particularly the cardiomyopathies.

Cardiac auscultation plays a limited role, but identifies individuals with aortic stenosis and approximately 25% of those with HCM. The 12-lead ECG is useful in the diagnosis of pre-excitation, congenital LQTS and Brugada syndromes. However, the classic ECG pattern of pseudo right bundle branch block and ST elevation in leads V1–V3 in Brugada syndrome may be transient, but can be unmasked by pharmacological provocation tests using sodium ion channel blockers (ajmaline or flecainide).[9] Such provocation tests are only reserved for athletes with a family history of the syndrome. The 12-lead ECG is non-specifically abnormal in up to 90% of individuals with heart muscle disorders; however, echocardiography is the most practical gold standard investigation. Visualisation of the origins of both coronary ostia on the short axis view of the aorta during two-dimensional echocardiography is a useful non-invasive method of diagnosing coronary artery anomalies. Subsequent imaging by coronary angiography or cardiac magnetic resonance will delineate the anomaly.[8] Exercise testing for objective assessment for myocardial ischaemia in patients with suspected coronary anomalies lacks sensitivity.[8]

Box 2 Sudden cardiac deaths in structurally normal hearts

- Congenital long QT syndrome
- Brugada syndrome
- Wolff–Parkinson–White syndrome
- Right ventricular outflow tachycardia
- Catecholaminergic polymorphic tachycardia
- Coronary vasospasms
- Commotio cordis
- Electrolyte disturbances
- Hypotherma or hyperthermia
- Drugs (amphetamine or cocaine)

Exercise stress testing is, however, useful to ascertain the broader aspects of the disease phenotype in suspected LQTS with a normal surface ECG and in the risk stratification of individuals with HCM. Athletes suspected to have ARVC usually require more detailed imaging with cardiac magnetic resonance, as fibro-fatty infiltration of the right ventricular wall is not well defined with two-dimentional echocardiography.[10]

In the past 15 years, there have been major advances in the molecular genetics of HCM, ARVC and congenital LQTS. However, the diagnosis of cardiovascular disease in athletes continues to rely predominantly on clinical investigations, as a genetic diagnosis is only possible in up to 60% of cases of HCM and an even smaller percentage in patients with ARVC and congenital LQTS.

DIFFERENTIATING ATHLETE'S HEART FROM HYPERTROPHIC CARDIOMYOPATHY

A small proportion of male endurance athletes develop substantial left ventricular hypertrophy (LVH; 12–15 mm) as part of a normal physiological adaptation to exercise which may mimic morphologically mild forms of HCM (8%).[11] The differentiation between physiological LVH and HCM is crucial, as an erroneous diagnosis can have potentially fatal consequences. In most cases, the differentiation is possible on the basis of family history, 12-lead ECG, left ventricular cavity size, indices of diastolic function and cardiopulmonary exercise gas exchange parameters.[12] In general, a positive family history of HCM, non-dilated left ventricular cavity, abnormal diastolic function and low peak oxygen consumption values favour HCM. In some cases, re-evaluation with ECG and echocardiogram after a short period of detraining to check for regression of physiological LVH may be the only practical method to distinguish between the two entities.[13]

MANAGEMENT

Definitive management of an athlete is dependent on the pathology identified. Most deaths are potentially triggered by physical exertion, therefore the most pragmatic approach in preventing such catastrophes would be to recommend abstinence from medium-intensity and high-intensity competitive sports.[14] Persuading a high-profile athlete to refrain from competitive sports can be difficult and sometimes impossible. Some athletes may accept the risk of SCD and continue to be involved in competitive sports in order to be in the athletic arena. The Bethesda guidelines actually recommend disqualification rather than advice to abstain from competitive sports[14]; however, decisions to disqualify cannot be legally enforced in the UK.

β-blockers are important in controlling symptoms in conditions such as HCM,[10] retarding aortic root dilatation in Marfan syndrome and preventing syncope and SCD in congenital

LQTS.[15] The prophylactic use of implantable intracardiac defibrillators is recommended in some patients with HCM, ARVC, congenital LQTS and Brugada syndrome who are considered to be at high risk of life-threatening ventricular tachyarrythmias.[7 9 10 15] Pre-excitation syndromes are best managed with radiofrequency ablation of the culprit electrical pathways. Athletes with the diagnosis of myocarditis are advised to abstain temporarily from competitive sports until their cardiac function is fully recovered.[14]

PREVENTION OF SCD IN ATHLETES

The identification of cardiac disorders in an athlete can prevent sudden death. A cost-effective pre-participation programme adopted by the USA that comprises a health questionnaire and physical examination is insensitive. Our own experience of screening athletes from various sporting disciplines suggests that the vast majority (>80%) of athletes identified with potentially serious cardiac disorders are completely asymptomatic. The Italian screening programme, which involves mandatory annual 12-lead ECG and a limited exercise test in all athletes, is associated with a higher identification rate of HCM but has major resource and cost implications. Currently, the most pragmatic approach in the UK would be to raise awareness of conditions causing SCD and to offer screening to athletes with symptoms highly suggestive of underlying cardiac disorder or a family history of inherited cardiac disorders and/or premature SCD. However, this should not deter more financially endowed sporting bodies from organising detailed cardiovascular screening for their young athletes before acceptance for competition.

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